SPINE RESEARCH

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Surgical management of spinal chordomas

A narrative review

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Abstract:
Chordomas are rare malignant tumors arising from remnants of the notochord, accounting for approximately 1% to 4% of all skeletal malignancies. These tumors are typically slow-growing and most commonly involve the sacrum, skull base, and spine. Their management is particularly challenging because of their proximity to critical neural and vascular structures. Although chordomas have a relatively low risk of distant metastasis, they are characterized by a high rate of local recurrence, necessitating aggressive, carefully planned treatment strategies. A narrative review of the literature was conducted to evaluate current approaches to the management of spinal chordomas, with a particular focus on surgical techniques and the role of adjuvant radiotherapy (RT). Relevant studies were identified through electronic database searches, including MEDLINE, Scopus, ProQuest, and Google Scholar. The selected literature was analyzed to assess surgical approaches, associated complications, oncological and functional outcomes, and the evolving role of RT in treatment. Surgical resection remains the cornerstone of treatment, with en bloc excision and negative margins as the gold standard. However, achieving complete resection, especially in the cervical spine, is often technically demanding due to anatomical constraints. For sacral chordomas, sacrectomy is frequently employed, while en bloc spondylectomy is considered for thoracolumbar lesions. Despite aggressive surgical strategies, complete tumor removal is not always feasible. In such cases, advancements in RT, particularly proton beam therapy and carbon ion therapy, have emerged as promising adjuncts. These modalities offer improved dose delivery while sparing surrounding critical structures. Nevertheless, the overall benefit of adjuvant RT remains a subject of ongoing debate in the literature. Postoperative complications are not uncommon and may include wound infections, neurological deficits, and mechanical instability, underscoring the complexity of managing these tumors. Given these challenges, a multidisciplinary approach involving neurosurgeons, orthopedic surgeons, radiation oncologists, and rehabilitation specialists is essential. In conclusion, the management of spinal chordomas requires an individualized and balanced approach that considers surgical feasibility, preservation of neurological function, and long-term disease control. While en bloc resection remains the mainstay of treatment, emerging RT techniques may enhance outcomes in selected patients. A coordinated multidisciplinary strategy is crucial to improving both prognosis and quality of life.

Abbreviations: ASIR = age-standardized incidence rate, BED = biologically effective dose, CT = computed tomography, C1–C6 = cervical vertebrae 1 to 6, DXA = dual-energy X-ray absorptiometry, GM = gluteus muscle, GTR = gross total resection, GyRBE = gray relative biological effectiveness, HR = hazard ratio, MRI = magnetic resonance imaging, PSM = paraspinous muscle, RT = radiotherapy, SBRT = stereotactic body radiation therapy, STR = subtotal resection, VRAM = vertical rectus abdominis myocutaneous, VSD = vacuum sealing drainage.

Keywords: chordomas, sacral chordomas, spinal chordomas, spinal tumors

1. Introduction

Chordoma is a primary bone sarcoma that is locally aggressive and originates from notochordal remains in the vertebral bodies along the spinal axis.[1] Virchow first characterized chordoma in 1846; it is an uncommon primary tumor with malignancy that makes up 1% to 4% of skeletal malignancies and develops from the embryonic remains of the notochord.[2,3] In the adult group, all forms of chordomas are more common in men.[4] Chordoma makes up 40% of primary spinal tumors and 2% to 4% of all spinal column tumors. The sacrum (50%), skull base (30%), and mobile spine (20%) are the locations of chordomas.[5] In mobile spine, they are mostly seen in the cervical spine. Patients with chordoma typically have a life expectancy of 6 years. Survival rates are 70% to 80% after 5 years and 40% to 60% after 10 years. Metastasis affects 30% to 40% of patients in advanced stages.[5,6] While spinal and sacral chordomas were most common in the Caucasian population, cranial chordomas were more common in Asian/Pacific people.[4] Chordomas may also

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Spine Research (2026) 2:2;101–109
Received: 12 October 2025 / Accepted: 14 February 2026

Published online 15 June 2026

http://dx.doi.org/10.1097/br9.0000000000000031

have an impact on young adults and children. In contrast to the craniocervical and sacrococcygeal prevalence in the adult populations, there is a female preponderance in the pediatric group, and it is more frequently appearing at the craniocervical junction with declining incidence caudally along the spine.[7] Given their placement along the midline of the axial skeleton and their close connection to the brainstem, cranial nerves, and spinal cord, chordomas have a poor natural course that is significantly accompanied by morbidity and death.[8] Chordoma makes up 4% of cancerous bone tumors, with an incidence of around 1 per 1 million individuals.[2,9] Chordomas are slow-growing tumors that have a low initial tendency for metastasis but a high probability of local relapse.[2]

In radiographic examinations, chordomas commonly show up as a degenerative lesion that damages the axial skeleton and is connected to a sizable soft tissue mass at the tumor's core within the vertebral body.[10] In contrast to osteosarcoma and chondrosarcoma of the vertebral body, which are more frequently detected in the appendicular bone, chordomas are malignancies of the soft tissue.[10] Chordomas are frequently present on computed tomography (CT) scans as an osteolytic, mixed osteolytic, and osteosclerotic bone lesion that typically contains a myxoid component.[10] Intravenous infusion of contrast mediates the visualization of chordomas.[10] Due to the more widespread use of magnetic resonance imaging (MRI), chordomas are now more frequently found in patients in their 30 s than in the past.[11] Chordomas exhibit hyperintense on MRI but can vary from isointense to hypointense on T1-weighted images.[10] Gadolinium contrast may be utilized as well to see improved images of the chordoma.[10]

The main course of treatment remains en bloc resection; however, some patients cannot have it because of the location and possible consequences.[1] Understanding chordomas has advanced; immunohistology markers like brachyury help make a positive diagnosis.[1]

Although radical excision at initial presentation has been advised for many years, it is not practical in many cervical spine cases; sacral tumors can grow to enormous sizes before diagnosis, and tumor-free margins are sometimes unattainable.[12] Some centers have started using proton therapy as the main treatment for chordomas because they are resistant to traditional radiotherapy (RT).[13–15] Thus, to discuss this contemplative topic further on, the authors aimed to review the literature regarding principles of surgical management, efficacy, survival rates, and complications of spinal chordoma surgery. Additionally, we also reviewed studies concerning the adjuvant RT given to chordoma patients.

2. Methods

To find articles about patients with spinal chordomas who underwent surgical intervention as well as RT, the authors searched the electronic literature. MEDLINE, Scopus, ProQuest, and Google Scholar were among the databases used. Keywords, "chordoma" [OR] "chordomas" [AND] "spine" [OR] "spinal" [OR] "sacrum" [OR] "lumbosacral" [OR] "lumbar" [OR] "thoracic" [OR] "cervical" [AND] "surgery" [OR] "radiosurgery" [OR] "radiotherapy" [OR] "radiation therapy" [OR] "proton" [OR] "photon" [OR] "carbon ion" [OR] "adjuvant therapy" [OR] "chemotherapy," were utilized as search phrases to conduct a literature search. Additionally, the retrieved papers' references were checked for other articles.

3. Results

3.1. Surgical management

The gold standard for treating spinal chordomas remains en bloc resection, which is the principal surgical procedure used. However, depending on the anatomical location and size of the tumor, the viability of obtaining total excision can vary greatly, requiring a comparison of different surgical approaches.

3.1.1. En bloc resection. This method has the best possibility of controlling the disease locally, especially when negative margins are possible. Research shows that this method reduces recurrence rates compared with piecemeal resection; however, there is a significant risk of intraoperative complications, especially with regard to neurological structures near the tumor.[16]

3.1.2. Piecemeal resection. Although less morbid than en bloc resection, piecemeal resection frequently results in greater rates of recurrence.[12] Patients who are not surgical candidates for en bloc resection may be candidates for this procedure, but it requires careful patient selection and rigorous follow-up to control the risk of recurrence.[17–19]

3.1.3. En bloc spondylectomy and sacrectomy. These methods have been developed especially for thoracolumbar and sacral chordomas, respectively.[12] There is evidence that these therapies can offer practical solutions for large tumors. The possible advantages of gross complete resection must be balanced against the considerable risk of problems associated with these surgeries, such as mechanical instability and postoperative infections.[11,20]

3.2. Surgical efficacy

The preferred treatment for cervical chordoma is total spondylectomy.[21] The conventional anterior reconstruction technique, which uses large autologous bone or titanium mesh bone graft in conjunction with titanium plate fixation, has good clinical results, but it also has drawbacks, including a limited supply of large autologous bone, a lengthy bone fusion time, and more frequent complications like internal implant displacement and titanium mesh sinking.[22] Additionally, it is challenging to locate an appropriate titanium mesh to reconstruct the stability of the cervical spine after vertebral excision of several segments, particularly tumors involving more than three segments. Furthermore, it is challenging to achieve dependable stability right away when reconstructing the cervical spine using titanium mesh because the anatomical morphology

of the cervical spine is unique, particularly the upper cervical spine, and the morphology of the titanium mesh differs greatly from that of the vertebral body.[23]

Advancements in surgical methods have also been achieved through artificial vertebral reconstruction. In contrast to conventional titanium mesh, three-dimensional (3D) printing allows for the customization of artificial vertebral bodies according to the angle of the adjacent vertebral endplates and the segments of the tumor. The use of 3D printed prosthetic vertebral bodies for repair following surgery for cervical spine tumors was primarily documented in case reports due to the brief clinical application of these structures.[24,25] A retrospective analysis of 24 patients with cervical chordoma who had total vertebral resection at Peking University Third Hospital between March 2005 and September 2019 included 15 patients undergoing titanium mesh reconstruction and 9 patients receiving artificial vertebral reconstruction using 3D printing.[26] Compared with titanium mesh reconstruction, 3D printed artificial vertebral bodies offered dependable immediate and medium-to-long-term cervical stability in total spondylectomy for cervical chordoma, lowering the risk of postoperative implant subsidence.[26]

In the evidence retrieved from the last 5 years, it was observed that surgical intervention plays a crucial part in enhancing survival rates.[5,27] Subtotal resection (STR) and gross total resection (GTR) patients had considerably lower hazard ratios.[27] Although malfunctions were also associated with anterior column repair and hardware malfunctions after en bloc removal of upper cervical chordoma.[28] For sacral and thoracolumbar chordomas, the best results were yielded with sacrectomy and en bloc spondylectomy.[12] The same study also found complications in mobile spine surgery.[12] Surgical procedure of 360° four-level en bloc showed good bone fusion with no recurrence, but there was a high rate of structural failure suspected.[29] Wang et al. studied that for lateral multilevel cervical chordoma, parasagittal osteotomy was a good alternative for en bloc spondylectomy.[16] One vertebral artery was preserved with the en bloc resection.[16] Yet, patients' nerve roots and vertebral arteries were sacrificed (see Table 1).

3.3. Anatomical region stratification

We found a great deal of variation in treatment strategies and results depending on anatomical regions.

3.3.1. Upper cervical region. En bloc resection in the upper cervical region is the intervention of choice; however, the proximity to important brain regions frequently makes it difficult to achieve distinct margins.

Table 1

Surgical efficacy for spinal chordoma.

Authors	Approach	Efficacy	Complications
Agner and Larkins[27]	•GTS (315 patients) •STR (308 patients)	➢Significant survival benefit with surgical intervention ➢Lower hazard ratios for STR and GTR ($p < 0.001$) vs. nonsurgical patients	○Poor outcomes with: -Increased age -Advanced disease stage -Chordoma of sacrum/pelvis vs. vertebrae -Absence of surgery
Patel et al.[5]	•Partial resection (499 patients) •Radical resection (550 patients)	➢Higher overall survival noted postsurgery	○Lower survival linked to: -Older age -Larger tumors -Presence of metastases
Pinter et al.[28]	•Free vascularized fibular graft after en bloc removal of high cervical chordoma (C1–C3)	➢Reports of negative margins; limited long-term outcome data	○Hardware malfunctions ○Complications from anterior column repair
Zileli and Karakoç[12]	•C2 chordoma: Anterolateral retropharyngeal excision Combined anterior–posterior approach Subaxial chordomas: Anterolateral corpectomy •Thoracolumbar chordomas: Total spondylectomy •Sacral chordomas: Sacrectomy	➢Sacrectomy gives the best results for sacral chordomas ➢En bloc spondylectomy is best for thoracolumbar chordomas	○Mobile spine surgeries have higher complication rates ○Sacral procedures lead to wound and urinary/rectal complications
Aoun et al.[29]	•360° four-level en bloc (C3–C6).	➢Bone fusion and no tumor recurrence reported	○High rate of structural failures ○Reoperations required
Wang et al.[16]	•Two-stage en bloc excision of multilayer cervical chordomas •Parasagittal osteotomy used	➢Preserves 1 vertebral artery while achieving en bloc status	○Sacrificed nerve roots in two patients ○Ligation of vertebral arteries in three patients

GTS, gross total resection; STR, subtotal resection.

According to the reviewed studies, full excision rates are significantly lower in this region than in others, and the complexity of these procedures is reflected in the reported postoperative consequences.[30–35]

3.3.2. Thoracolumbar region. For thoracolumbar chordomas, en bloc spondylectomy is frequently employed. The literature suggests that larger tumor sizes correlate with increased surgical difficulty and a higher incidence of local recurrence.[12] This substantiates the need for tailored surgical strategies in this anatomical section.

3.3.3. Sacral region. Sacrectomy presents unique challenges, especially concerning preservation of neurological function and pelvic stability.[36,37] Our review indicates that sacral chordomas have different recurrence patterns and complications compared with those found in the cervical and thoracolumbar regions, correlating with their anatomical and functional implications.

3.4. Tumor characteristics

Tumor size has a major impact on surgical results and how feasible it is to achieve negative margins. According to published research, tumors larger than a particular threshold are associated with increased chances of complications and recurrence.[5] Vascular involvement makes surgical resection more difficult, especially in bigger tumors, requiring meticulous preoperative planning and interdisciplinary intervention.[33]

3.5. Reconstruction after removal of chordoma

A case investigation revealed a lobulated extradural mass that spanned from C3 to C6, severely compressing the spinal cord and damaging the vertebral body.[38] The procedure was carried out using a sequential circumferential approach. Anterior corpectomy and tumor removal with reconstruction utilizing an anterior plate and titanium cage comprised the first step; posterior instrumentation for stabilization made up the second. A histopathological analysis verified the presence of brachyury expression in typical chordoma. After 2 months, the patient had a significant neurological recovery and was recommended for adjuvant RT.[38] The case report by Pinter et al. documented reconstruction using a free vascularized fibular graft after an en bloc removal of a chordoma affecting C1–C3.[28] This report was the first to show satisfactory long-term oncological results following true margin-negative excision of a high cervical chordoma. It also showed the construct's sustainability at a 10-year follow-up.[23]

Following sacral chordoma excision, patients have four dependable options: free flaps, transpelvic vertical rectus abdominis myocutaneous (VRAM) flaps, bilateral gluteal advancement flaps, and direct closure.[37] Rajagopalan et al. conducted a retrospective analysis of patients who had reconstruction following the excision of lumbar or sacral chordomas at a single facility between 2012 and 2023.[39] The majority of the patients received gluteus muscle flaps. The second most frequent flaps were those of the paraspinous muscle (PSM) and VRAM. Hardware placement was more frequently linked to VRAM and PSM flap reconstruction ($p < 0.01$). The median follow-up period was 34 months. The incidence of postoperative surgical complications was not correlated with the method of reconstruction or the location of the hardware.[39] Two cases of significant deformities after total sacrectomy for chordoma were reported by Xu et al.[36] A modified lumbar artery perforator flap was used to treat case 1, a 65-year-old woman with a deep three-dimensional cavity, after the initial standard vacuum sealing drainage (VSD) was unable to eliminate the dead space. In case 2, a 45-year-old man with a significant surface defect had conservative treatment using a modified VSD technique in conjunction with silver ion dressings. In case 1, full and long-lasting wound healing resulted from the perforator flap's solid, vascularized covering. In case 2, a silver dressing regimen and improved VSD effectively prevented possible infection and encouraged strong granulation, leading to full wound closure by secondary intention. Both patients had well-healed wounds during follow-up, showing no signs of serious consequences or recurrence.[36] For postsacrectomy deformities, the aforementioned report emphasizes that a single reconstructive technique is not always appropriate. From expert conservative care with modified VSD and antimicrobial dressings to intricate perforator flap restoration, a graduated and customized strategy is crucial. However, for patients with partial sacrectomy, intact gluteal vessels, and no preoperative RT, bilateral gluteal advancement flaps or gluteal perforator flaps are the first option.[37] For patients with near-total sacrectomy and preoperative RT, transpelvic VRAM flaps or free flaps are the next options.[37]

3.6. Complications

3.6.1. Cervical chordomas. Many researchers contend that an aggressive surgical approach is constrained by the potential of neurovascular damage since cervical spine chordomas typically include important anatomy like the vertebral arteries and cervical nerve roots.[30–33] Because of the diversity of the radiculomedullary branches, vertebral artery sacrifice can result in spinal cord ischemia and stroke, especially if the tumor involves the dominant vertebral artery.[33,34] To assess the viability of vascular sacrifice for en bloc excision, a number of investigators have suggested that patients with chordomas involving the vertebrae undergo a cerebral angiography and a transient balloon occlusion test as part of the preoperative workup.[33] Since doing so results in a neurological deficit (loss of function in the upper limb muscles), which is crippling and lowers quality of life, scholars have been hesitant to sacrifice nerve roots in subsequent experiments.[33] En bloc advocates suggest that if long-term survival or a possible cure are enhanced, neurological deficits would be tolerable.[33] It is interesting to note that no notable neurological impairments were

observed in patients who had the C1–C6 cervical nerve root(s) excised.[32,33,40]

3.6.2. Sacral chordomas. An early scar disunion following surgery necessitates surgical revision and ongoing antimicrobial therapy.[11] Additionally, it's possible that it is a late infection.[11] When compared with laparotomy, the anterior technique requires less blood loss.[41,42] Nevertheless, the primary source of bleeding is the posterior approach with the sacral osteotomy.[11] Even without RT, sacrectomy above the S2 carries a risk of pathological fracture; hence, prophylactic internal fixation should be used.[11]

4. Discussion

4.1. Outcomes of chordoma surgery

4.1.1. Metastasis, recurrence, and survival. After broad excision, there is a 40% to 20% local recurrence rate of sacral chordoma and a 20% to 40% metastatic recurrence rate.[43] Difficulty in performing successful wide resection or GTR of the spinal chordomas is demonstrated by high rates of recurrence and metastasis, which emphasizes the significance of cautious surgical management of these patients.[44] Because infection in the resection margins can increase the risk of recurrence and mortality, as shown in a meta-analysis, it is crucial to achieve a wide excision.[45] A database analysis has shown not much benefit in cases where GTR has been achieved.[46] Promising developments have been made in more accurate RT techniques, such as stereotactic RT, carbon ion treatment, and proton beam therapy.[44]

There have been some advances in chordoma survival with recent therapeutic techniques. Surgical margins have little effect on results, despite the fact that numerous earlier studies have supported negative surgical margins, also referred to as Enneking negative surgery.[1] Recent developments in diagnostic and therapeutic approaches, such as improved tumor resection techniques and improved RT methods like proton beam therapy, may have contributed to improvements in the survival times of sacral and mobile spine chordomas.[47] Proton beam therapy is one example of a modern radiation technique that offers longer survival times and fewer complications than surgery.[1] Table 2 displays the outcomes of the discussed interventions in detail.

Usually, metastases develop in the bones, subcutaneous tissues, lymph nodes, and lungs.[12] Following a recurrence, metastases are more commonly seen.[12] Compared with chordomas of the mobile spine, sacral chordomas have a higher propensity to spread.[12]

Table 2

Literature on survival/recurrence rate of spinal chordoma in the last 5 years.

Authors	Year	Study type/level of evidence	Number of patients/ articles	Intervention	Survival/recurrence rate
Agner and Larkins[27]	2024	Retrospective study (surveillance, epidemiology, and end results program)/3	896 patients	Surgery, Conventional RT, Photon RT, Carbon ion RT	A 5-year overall increase in survival was noticed. STR HR: 0.48 GTR HR: 0.22 RT HR: 0.69
Patel et al.[5]	2023	Retrospective study (National Cancer Database)/3	1,548 patients	Surgery, Conventional RT, Photon RT, Carbon ion RT	Mean overall survival: 8.2 years Partial resection HR: 0.43 Radical resection HR: 0.35 RT HR: 0.82
Pennington et al.[47]	2021	Narrative review/4		Surgery, Conventional RT, Photon RT, Carbon ion RT	High-dose (> 50 Gy) neoadjuvant or adjuvant RT is becoming the standard of care because of advancements in radiation modalities (photon therapy, proton beam radiation).
Zileli and Karakoç[12]	2021	Retrospective clinical series (6-year follow-up)/3	48 patients	Surgery, Conventional RT	**Sacrectomy:** Average survival period: 68 months Recurrence rate: 76% **Marginal or piecemeal resection:** Average survival period: 76.5 months Recurrence rate: 83%
Baig et al.[44]	2021	Systematic review/3	1,531 patients (42 articles)	Surgery, Conventional RT	GTR recurrence rate: 25% STR recurrence rate: 50%
Chehrassan et al.[48]	2020	Retrospective study (National population-based study)/3	122 patients	Not investigated	Mean survival time: 4.5 years. Age-standardized incidence rate (ASIR) of chordoma: 0.28. Sacral chordoma cases: 67.2%
Denaro et al.[2]	2020	Systematic review/3	1,359 patients (58 articles)	Surgery, Conventional RT, Photon RT, Carbon ion RT	Cervical chordomas recurrence rate range: 25%–60% Sacrococcygeal chordomas recurrence rate range: 18%–89%

GTS, gross total resection; HR, hazard ratio; RT, radiotherapy; STR, subtotal resection.

4.1.2. Feasibility. Anatomical limitations make it difficult to achieve total excision through en bloc resection. It highlights how crucial it is to take surgical viability into account when creating a customized treatment plan for patients, weighing the possibility of effective tumor removal against functional preservation and overall disease control.[49] When choosing the optimal course of therapy, this feasibility evaluation is essential.

4.1.3. Morbidity trade-off. Due to their proximity to important neural structures such as the brainstem, cranial nerves, and spinal cord, chordomas frequently have a poor natural course, resulting in considerable morbidity and the possibility of death.[8]

Although en bloc resection is the primary treatment strategy to achieve negative margins, a number of circumstances, including the anatomical limitations of the tumor's placement, might make this technique challenging. Postoperative problems include infections, neurological impairments, and mechanical instability. These complications may arise from the challenging resection.[33]

To maximize patient care, a multidisciplinary approach is required, weighing the possible advantages of illness control and functional preservation against the dangers of surgery (morbidity).

4.2. RT in spinal chordoma

When total surgical excision is not practical, RT is an additional treatment option for spinal chordomas. Each of the offered modalities has distinct benefits and drawbacks that call for careful consideration (see Table 2).

4.2.1. Proton beam therapy. This technique is preferred due to its ability to target tumors while protecting nearby healthy tissues precisely. When paired with surgery, recent research indicates better results in terms of recurrence.[49] The best dosage guidelines and long-term effects, however, are still contentious topics.

4.2.2. Carbon ion therapy. A powerful substitute, carbon ion therapy has advantages over traditional forms of radiation because of its higher linear energy transfer, which could result in better tumor control. Although the first results are encouraging, similar to proton treatment, integrating it into conventional care is difficult due to the lack of comprehensive comparative data and conclusive protocols.[50]

4.2.3. Traditional photon treatment. Although photon treatment is widely available, its efficacy in treating spinal chordomas is questionable since it may cause harm to nearby structures, which could raise long-term morbidity.[2,5,27,47]

4.3. Dose–response relationships

Research repeatedly demonstrates that dose escalation is essential for attaining local control in sacral and mobile spine chordomas. Studies on proton beam therapy show better results when total doses are above 70 gray relative biological effectiveness (GyRBE), especially in patients with positive or close margins.[51] Similarly, when ablative doses are administered, such as 24 Gy in a single fraction or biologically effective doses (BED2) larger than 140 Gy, stereotactic body radiation treatment (SBRT) exhibits remarkable control.[52] Although there remains a lack of data relevant to the spine, carbon ion therapy's greater relative biological effectiveness further reinforces the case for dose escalation.[53]

4.4. Comparative efficacy

Comparisons between modalities indicate that proton treatment offers better local control than photon therapy due to its improved conformality and ability to spare nearby organs. With long-term studies in skull base chordomas demonstrating local control rates of 70% to 76% at 5 years, carbon ion therapy gives even better biological efficacy.[54] With retrospective data demonstrating local control rates of up to 95% in treatment-naive patients, SBRT has become a popular choice.[53] Although surgery remains the mainstay of treatment, patients who are medically inoperable can be reasonably controlled with radiation alone.

4.5. Limitations

The majority of retrospective, single-institution series with few patients and a variety of treatment modalities constrain the literature. Given the lengthy natural history of chordomas, it is troublesome that follow-up periods are frequently brief. Treatment planning is made more difficult by technical issues such as dosimetric ambiguity brought on by surgical devices.[55,56] Different modalities have different toxicities. Proton or carbon ion therapy is more likely to cause problems with wound healing, but SBRT typically has lower rates of skin toxicity.[53] The majority of recommendations are based on expert consensus rather than high-level evidence due to the lack of randomized controlled trials.

4.6. Current controversies

There are still a number of unsolved disputes. Neoadjuvant therapy offers sharper target delineation but raises issues regarding wound healing[57]; the timing of RT,[58] whether adjuvant or neoadjuvant, remains up for dispute. Another point of contention is the determination of target volumes: focal volumes run the danger of marginal failures, whereas comprehensive sizes enhance local control but raise toxicity.[53] Due to a lack of comparison data, the choice of modality, for example, proton, carbon ion, SBRT, or combination photon/proton plans, remains institution dependent. Finally, even though major surgery and high-dose radiation can improve local control, striking a balance between quality of life and aggressive therapy is a constant issue.

5. Conclusion

There are significant difficulties in managing spinal chordomas, especially when it comes to metastases and recurrences. Sacral chordomas and mobile spine chordomas have similar survival and recurrence rates. Mobile spine chordomas do not typically progress to metastases, while sacral chordomas do. For sacral chordomas at S2 and lower, sacrectomy is effective. Unfortunately, additional marginal excision of cervical chordomas is not feasible without serious impairments and problems. The main treatment for chordoma should be surgical resection. Although drastic surgery is frequently highlighted, the viability should be assessed on an individual basis using a multidisciplinary approach customized to meet the needs of each patient.

The patient should be carefully informed about the surgical process, expected advantages, and any problems. It is difficult to standardize this operation, and the surgeon must know how to modify the surgical process to meet the needs of each patient. As a result, the surgical technique and fixing method must be uniquely tailored to the individual patient. The patient should be psychologically and physically prepared, as well as informed about the difficulties and anticipated aftereffects that may reduce the quality of life. If a colostomy is anticipated, learning how to self-catheterize may be necessary before surgery, along with visiting a stoma therapist. Thus, surgical management requires multidisciplinary skills from the fields of surgery, anesthesia, radiology, and so on. Surgery carried out outside of a referral facility is a separate risk factor for fatal results.

Acknowledgements

Not applicable.

Ethical statement

Not applicable.

Conflicts of interest

The authors have no conflicts of interest to disclose.

Funding source

This research received no specific grant from any funding agency in the public, commercial, or not-for-profit sectors.

Data availability statement

Data sharing not applicable to this article as no datasets were generated or analyzed during the current study.

Author contributions

UY—conceptualization, methodology, data curation, formal analysis, and drafting of the manuscript.
FZ—methodology, and critical revision of the manuscript.
AAK—data curation, formal analysis, and manuscript revision.
SMH—investigation, visualization, and review of the manuscript.
FZa—data curation and manuscript revision.
TTA—supervised the project, contributed to project administration, and critically reviewed the manuscript.
All authors read and approved the final manuscript.

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How to cite this article: Yaqoob U, Zahid F, Khan AA, Hussain SM, Zahra F, Awan TT. Surgical management of spinal chordomas: A narrative review. *Spine Res*. 2026;2(2):101–109. doi: 10.1097/br9.0000000000000031